**FAMILIAL MITOCHONDRIAL CARDIOMYOPATHY**

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Mitochondrial disorders are disorders due to impaired mitochondrial energy metabolism. Respiratory chain deficiency can be found in over 70% of patients with mitochondrial disease, where cytochrome *c*oxidase deficiency may lead to overall complex I deficiency, and can account for up to a third of the cases. The majority of oxidative phosphorylation diseases identified thus far are caused by mitochondrial DNA mutations and are maternally-inherited. Mitochondrial cardiomyopathy may present as part of a neurologic disorder, such as MELAS, MERFF, Kearns-Sayre or Leigh syndromes, or may manifest as isolated fatal infantile cardiomyopathy. Since the heart, as well as the brain and the muscle, is mainly dependent on aerobic respiration for its energy requirement, the heart is one of the organs most frequently affected in mitochondrial disorders. The most frequent cardiac manifestation of a mitochondrial disorder is cardiomyopathy, of which the hypertrophic form is the most common. Dilated cardiomyopathy and left ventricular non-compaction may also be observed. Two cases will be presented as a part of my talk. Two siblings who exhibited progressive hypertrophic cardiomyopathy. Both appeared healthy at birth, without evidence of cardiac disease. Both became symptomatic at the age of 6 months with rapidly deteriorating combined cardiomyopathy (mostly hypertrophic), and both expired within a few months after clinical diagnosis was done and confirmed.